Respiratory Medicine (1996) **90,** 53–55

Case Reports

Recurrent respiratory papillomatosis with malignant transformation in a young adult

D. Orphanidou, K. Dimakou, P. Latsi, M. Gaga, M. Toumbis, A. Rasidakis and J. Jordanoglou

Respiratory Medicine Department, School of Medicine, Athens University, Greece

Introduction

The term 'papilloma' was first used by Mackenzie 100 years ago, who claimed that this was the most benign tumour of the larynx (1). Today papillomas are considered to be caused by the Human Papilloma Virus group (H.P.V.) (2–6). The majority of patients suffering from this disease which is also referred to as 'recurrent respiratory papillomatosis' (2,7) require multiple surgical operations for tumour removal (8). Malignant transformation of papillomas, which is a rare condition, is considered to occur mainly to irradiated patients (5).

The following report describes the case of a male patient, with a history of vocal cord papillomas since his first year of age, who developed bronchial and pulmonary spread of the disease. He died at the age of 26 years because of squamous cell carcinoma which was related to the malignant transformation of the pulmonary papillomas.

Case Report

The patient was a 26-year-old male, non-smoker, builder in profession, with a history of vocal cord papillomas since the age of 1 year. At this early age, surgical excision of papillomas was performed and at the age of 2 years a tracheostomy was performed. His general condition was good until the age of 20 years, when the patient underwent electric cauterization of the papillomas which subsequently relapsed again. Following this, the patient had frequent respiratory infections, sometimes accompanied with haemoptysis. At the age of 25, he was first admitted to a respiratory unit because of low grade fever for 6 months and a few days duration of pleuritic chest pain. The chest X-ray showed a left pleural effusion which proved to be exudate with predominance of polymorphonuclear white blood cells. In addition, the chest computed tomography revealed bilateral small cavitating nodules. The patient improved clinically on antimicrobial therapy. Seven months later, he was admitted to our department with fever, cough with purulent expectoration, bloody sputum and cervical pain. Physical examination showed a young patient with kyphoscoliosis and tracheostomy. Ausculation of the thorax revealed bilateral inspiratory crackles over the middle and bases of both lungs. The findings of the remainder of the examination were within normal limits.

Chest X-ray showed a shadow in the left mid zone and left lower lobe atelectasis (Plate 1). Fibre-optic bronchoscopy was then performed which demonstrated polypoidal protrusions of the mucosa in the left lower lobar bronchus, that caused obstruction. Pathological examination of the endobronchial biopsies showed the presence of papillomas. Cytological examination of the bronchial washing and brushing were negative. High-resolution computed tomography of the thorax revealed multiple bilateral pulmonary solid and cavitating nodules in addition to the X-ray findings (Plate 2). Bone scanning showed increased radioactivity at the cervical part of the spinal column.

After control of the infection, laser treatment was recommended in order to widen the lumen of the left lower bronchus, but the patient refused.

Two months later, he was admitted to a hospital again because of increasing cervical pain and profound oedema of the neck. A new bronchoscopy was then performed. Cytological examination of the bronchial secretions, as well as the material obtained by percutaneous needle aspiration of the left peripheral solid nodule under ultrasound control, revealed the presence of squamous cell carcinoma.

Received 26 January 1994 and accepted 3 February 1994.

0954-6111/96/010053+03 $12.00/0

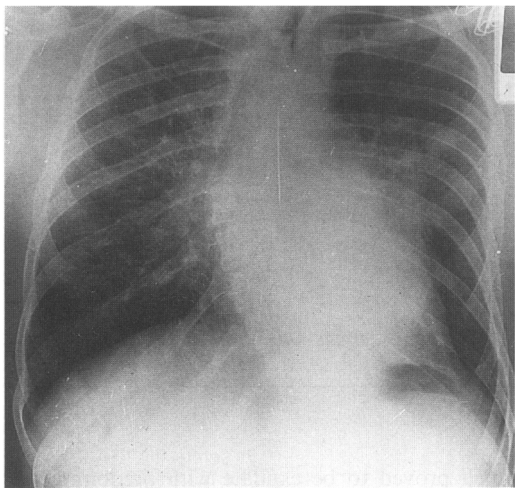

Plate 1 Chest X-ray showing a shadow in the left mid zone.

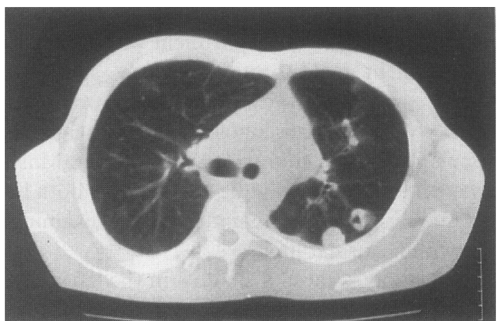

Plate 2 CT scan of the thorax showing bilateral cavitating nodules.

A few days later the patient developed paraplegia, his condition deteriorated rapidly and he died.

Discussion

The term recurrent respiratory papillomatosis (R.R.P) was first used in 1976 (9) to describe the development of papillomas in the upper or lower respiratory tract. More often, they appear in the larynx of children between 18 months and 3 years of age, where they remain restricted in the majority of cases (10). The trachea and bronchi are involved in about 5% of patients (4) and the lung parenchyma in less then 1% of patients (11). The development of papillomas is commonly associated with severe symptoms of obstruction, which often lead to an incorrect diagnosis (8).

Lung lesions do not tend to regress spontaneously while uncomplicated laryngeal papillomas often do (12). These lesions may occur in childhood (12–14) or adult life (12) as in the case of our patient in whom the lung lesions appeared 24 yr after their initial location in the vocal cords. The pathogenesis of their development in the lower respiratory tract is unknown. Various theories have been suggested such as the multicentric change of respiratory epithelium by the virus, or spread of tumour fragments during the endoscopic procedures (11,15,16).

The management of recurrent respiratory papillomatosis remains a perplexity. Treatment is palliative, the papillomas being removed using the carbon dioxide laser and/or cupped forceps (8). A limited number of case reports refer to the management of the disease with the use of photodynamic therapy (15). The frequent relapses of the tumours and the severe symptoms they produce cause a great problem. (8). Our patient who had tracheostomy since the age of 2 years did not develop a severe relapse of the disease, and did not, therefore, require any kind of treatment up to the age of 25 years.

Apart from operations for tumour removal, several methods of adjunctive treatment have been used occasionally as the administration of bleomycin (14), maternal transfer factor (13), poly (I,C) -LC (2) and interferon (17).

Although the disease is benign, a very small number of patients develop carcinoma as a result of the malignant transformation.

An analysis of a series of 113 patients with laryngeal papillomas showed that irradiated patients had a 16-fold increased risk of subsequent carcinoma in the respiratory system (5). There is, however, a small number of patients developing carcinomas who had not been irradiated, as in the case of our patient (5, 12). The case of our patient is presented because of the following three interesting points: (a) the disease was stable for a long period of time; (b) laryngeal, endobronchial and pulmonary papillomas were coexisting; and (c) malignant transformation of the papillomas.

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